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CASE REPORT

# Treatment options for transposition of the great arteries with ventricular septal defect complicated by pulmonary vascular obstructive disease

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**Abstract** The arterial switch operation is the procedure of choice for patients with isolated transposition of the great arteries or those with associated atrial and/or ventricular septal defects. After the development of pulmonary arterial hypertension, the surgical options for patients with a late presentation include either retraining the left ventricle by pulmonary artery banding followed by an arterial switch operation or palliative atrial or arterial switch, with or without medical management of pulmonary hypertension. We present a case with D-transposition of the great arteries with ventricular septal defects and irreversible pulmonary arterial hypertension who improved after a palliative atrial switch operation.



## 1. Introduction

Complete transposition of the great arteries (D-TGA) represents approximately 5–8% of congenital cardiac malformations but accounts for 25% of deaths from congenital heart disease in the first year of life (Liebman et al., 1969). Survival is highly dependent on the interplay and mixing between the two parallel circuits and the amount of pulmonary blood flow (Maeno et al., 2000).

The prognosis for unoperated patients with TGA with intact ventricular septum is poor. The overall mortality rate for unoperated cases is 29% in the first week, 52% in the first month, and 89% in the first year (Galie et al., 2004). Unoperated TGA associated with ASD and/or VSD has a

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better survival rate. The average life expectancy for patients with atrial or ventricular septal defect in addition to TGA is 9 or 22 months, respectively (Galie et al., 2004). At present, the typical treatment is a definitive corrective procedure (the arterial switch operation) early in infancy; with this procedure, many patients reach adulthood (Wijesekera et al., 2005) and they may have normal life expectancy.

## 2. Case report

We report an 8-year-old Saudi female child delivered vaginally at home after an uneventful full-term pregnancy. At 3 months of age, the infant was noticed to have bluish discoloration, diaphoresis, and excessive crying. She was diagnosed with TGA and a VSD. An atrial septostomy was performed, and she was referred for an arterial switch operation. Unfortunately, the parents misunderstood (or ignored) the plan and missed the referral and the follow-up visits. The child presented at 18 months of age with evidence of severe pulmonary arterial hypertension (PAH) due to pulmonary vascular disease and was declared inoperable.

She was referred to our centre at 8 years of age with severe exertional dyspnoea and exercise intolerance (NYHA functional class IV). On examination, she was physically well developed with a weight of 16.3 kg (below the 5th percentile) and a height of 120 cm (between the 10th and 25th percentiles). She was cyanosed with marked digital clubbing. She had physical signs consistent with right ventricular hypertrophy and clinical evidence of PAH. No signs of heart failure were evident. Her resting oxygen saturation was around 60%. Her haemoglobin was 18.5 g/dl, and haematocrit was 58 L/l. Transthoracic echocardiography demonstrated atrioventricular concordance, ventriculoarterial discordance and a large perimembranous VSD as well as a patent ductus arteriosus with severe tricuspid insufficiency. After a joint medical/surgical discussion, we considered her for a palliative atrial or arterial switch surgery. Diagnostic cardiac catheterisation was performed and revealed the presence of severe PAH with suprasystemic pulmonary arterial pressures. Her pulmonary artery saturation on room air was 91%, and the indexed pulmonary vascular resistance was 10 Woods units $m^2$. With 100% oxygen and inhaled nitric oxide at 20 parts per million for 10 min, the pulmonary arterial saturation rose to 97%; however, there was minimal change in the pulmonary vascular resistance. The decision was to proceed to atrial switch operation.

The patient underwent an atrial switch operation (Mustard procedure) and patent ductus arteriosus ligation with tricuspid valve repair. The VSD was not closed. The postoperative course was uneventful. Seventeen months post-procedure, she was NYHA functional class I, her resting saturations varied from 85% to 90%, her haemoglobin was 13.5 g/dl, and her haematocrit was 42 L/l. There were no signs of heart failure.

Her medications included furosemide 10 mg orally twice daily, captopril 1.5 mg orally three times daily, aspirin 50 mg orally once daily, and sildenafil 3 mg orally three times daily. Sildenafil was stopped 14 months post-operatively (to avoid an increase in pulmonary blood flow and the possible development of heart failure). No further surgical intervention was planned.

## 3. Discussion

Despite advances in cardiac surgery, systemic or suprasystemic pulmonary vascular pressure associated with congenital heart disease remains a major problem. Recently, advanced therapies for PAH have become available and are effective at reducing pulmonary vascular resistance and symptoms in patients with near-systemic pulmonary arterial pressures. Previously, these patients were thought to have irreversible pulmonary vascular disease. This raises an interesting question; specifically, can intra-cardiac communications previously considered inoperable due to severe pulmonary vascular disease become amenable to surgery after successful treatment with advanced therapy (Dimopoulos et al., 2008)?

The Eisenmenger syndrome is at the extreme end of the spectrum of PAH secondary to congenital heart disease. It includes all left-to-right shunts that result in significant systemic or near-systemic PAH with subsequent reversal of the shunt direction. It results in a significant increase in morbidity and mortality and greatly affects the quality of life.

The current definition of PAH relies on the presence of a mean pulmonary arterial pressure exceeding 25 mm Hg at rest or 30 mm Hg during exercise and a resting pulmonary vascular resistance above 3 Woods units (Galie et al., 2004; Rubin and Badesch, 2005). An estimated 5–10% of all patients with CHD, mostly those with late or no repair, have some form of PAH (Galie et al., 2004; Engelfriet et al., 2007).

In theory, all large communications that result in pulmonary over-circulation by means of a significant left-to-right shunt can produce progressive histologic and patho-physiologic abnormalities in the pulmonary vascular bed leading to PAH. VSDs are the most common simple defects causing PAH. An estimated 10% of all VSDs and 50% of large VSDs have the potential to cause Eisenmenger syndrome, if not repaired by age two (Hoffman and Rudolph, 1965).

An acute reduction in the mean pulmonary arterial pressure of 10 mm Hg with a resultant mean pulmonary arterial pressure of 40 mm Hg or less without a fall in cardiac output is considered a positive vasoreactivity response. Balloon test occlusion of the cardiac defect may provide additional information on the suitability for closure and the possible post-procedural outcome. A drop in cardiac output and/or an increase in right ventricular filling pressures with a test balloon occlusion would suggest a low likelihood of benefit from permanent closure as well as a higher perioperative risk (Allen et al., 2008).

Palliative can be done to reduce the hypoxia in patients with PAH secondary to TGA/VSD. In these patients the pulmonary arterial oxygen saturations is higher than that in the aorta (Bernhard et al., 1976; Burkhart et al., 2004). The increase in arterial oxygen saturation resulting from the redirection of flow provides significant symptomatic relief and increased quality of life in patients with severe cyanosis-related symptoms.

The goals of cardiac surgery in adult patients with CHD and PAH are to avoid complications of Eisenmenger syndrome, such as addressing the right-to-left shunt, decreasing cerebrovascular events (stroke and abscess), preventing cyanosis and improving exercise capacity, reducing erythrocytosis and haemostatic complications, reducing the incidence of systemic organ failure, and protecting the pulmonary circulation. Currently, no data are available on the long-term response of the right ventricle to closure of intra-cardiac communications

in patients with right-to-left shunting. Once the defect is closed, a patho-physiologic situation more similar to idiopathic PAH could become established, which is associated with a much worse long-term outcome than Eisenmenger syndrome (Dimopoulos et al., 2008).

In 1982, Jatene described the first successful arterial switch operation (Jatene et al., 1982). In 1987, Senning wrote: "I think that the arterial switch—a real anatomical correction—will be the 'gold standard' in the near future and the atrial switch will be used only for the few patients who are not suitable candidates for the arterial switch" (Bove, 1987).

The current indications for an atrial switch procedure include:

(1) Infants with isolated TGA (with intact ventricular septum) who present after the neonatal period. The alternative approach is pulmonary artery banding to train the left ventricle followed by arterial switch.
(2) Palliation of patients with pulmonary vascular disease from an associated ventricular septal defect (Burkhart et al., 2004).
(3) Patients with a congenitally corrected transposition in whom repair includes both a venous and arterial switch to create ventriculoarterial concordance or in patients with associated pulmonary valve stenosis or atresia, an atrial switch combined with a Rastelli operation (namely, Ilbawi's operation, also known as a "double switch" or "anatomical correction") (Ilbawi et al., 1990; Konstantinov and Williams, 2003).
(4) For the rare anomaly of isolated ventricular inversion, there is atrioventricular discordance with ventriculoarterial concordance (Pasquini et al., 1988). The atrial switch operation corrects the circulation physiologically and leaves the morphologic left ventricle supporting the systemic circulation. In this case, the arterial switch operation is contraindicated because of the presence of ventriculoarterial concordance.
(5) Patients with other complex congenital lesions with VSD, transposition haemodynamics, and severe pulmonary vascular obstructive disease (PVOD) (Current case) (Bernhard et al., 1976; Lindesmith et al., 1972; Corno et al., 1987; Konstantinov et al., 2004).

In the current era, newborns with transposition have a 20-year survival well over 95% after arterial switch operation (Williams, 2006).

In 1972, Lindesmith et al. were the first to report a series of patients with TGA, VSD, and severe PVOD who underwent a palliative atrial switch operation (PAS) (Lindesmith et al., 1972). This operation consisted of a Mustard procedure to reroute pulmonary and systemic venous drainage while leaving the VSD open. The VSD was not closed because experience indicated that closure in such patients was associated with significant early and late mortality (Mair et al., 1976). Their results, as well as those of others, have shown improved early mortality and considerable relief of symptoms in deeply cyanotic patients with severe PVOD (Mair et al., 1976; Sagin-Saylam and Somerville, 1996). A careful cardiac catheterisation assessment is essential for determining whether any patient should be considered for PAS.

Bukhart et al. reported a series of 24 patients with different pathologies who underwent a PAS. If the patient was only 1 or 2 years old and there was a good reduction in PVR with 100% oxygen inhalation, an atrial switch with VSD closure was considered. Alternatively, if the patient was older or had a poor response to 100% oxygen, the authors favoured PAS. If the VSD was closed in a borderline atrial switch candidate and the patient's haemodynamics were suboptimal after weaning from cardiopulmonary bypass, the bypass was reinstituted and a hole placed in the VSD patch. Pulmonary vascular resistance less than 8 Woods units $m^2$ may result in an unacceptable left-to-right shunt and ventricular failure after PAS. The authors reported a significant improvement in NYHA functional class and a survival rate (54%) after 15 years of follow-up (Burkhart et al., 2004).

The arterial switch procedure has become the operation of choice during the neonatal period for management of TGA with or without VSD. Case reports of its application (leaving the VSD open) in the setting of late presentation pulmonary hypertension and PVOD exist in the literature (Pridjian et al., 1992; Elizari and Somerville, 1999). A palliative arterial switch has some potential advantages over PAS, especially with respect to avoiding late atrial arrhythmias and baffle obstruction; however, long-term results in this setting are not yet available. Both procedures are valid and effective; at the present time, the decision regarding which surgery to perform is made according to the individual surgeon's experience and preference (Burkhart et al., 2004).

In summary, we report a patient with TGA and a large VSD with Eisenmenger syndrome who underwent a successful palliative atrial switch operation with improvement in functional capacity.

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